CLINICAL CASES

Reported from the Clinic of Prof. M. Allen Starr, College of Physicians and Surgeons, New York.

FRIEDREICH'S ATAXIA.

This disease is sufficiently rare to warrant the report of cases.

Case I. Susan J., aged thirteen in December, 1897, is the second of five children. Her father and mother are healthy, and no one of her grand parents, aunts or uncles are known to have had any form of paralysis. An older brother, however, suffered from this disease which developed at the age of eleven and rendered him a cripple until his death from diphtheria last year. The patient was a healthy child, though it was noticed that as a baby, while learning to walk, she had a somewhat waddling gait and was clumsy on her feet. Her present illness, however, did not begin until the age of eight, after an attack of measles. Since that time difficulty in walking has gradually and progressively increased, and one year after the beginning of the trouble in the legs it was noticed that the hands were used in a clumsy manner. During the past five years she has suffered some from pains which were supposed to be rheumatic, and has occasional retention of urine which has never, however, required the use of a catheter. She applied at the clinic on account of the unsteadiness of her gait in walking, and examination showed her gait to be markedly ataxic, the steps being irregular in length, the feet being placed too widely apart, but tending to overlap in walking unless corrected by voluntary movements to preserve her equilibrium, the feet dropping somewhat with the toes turning inward and, hence, being lifted too high from the floor in the act of walking. It

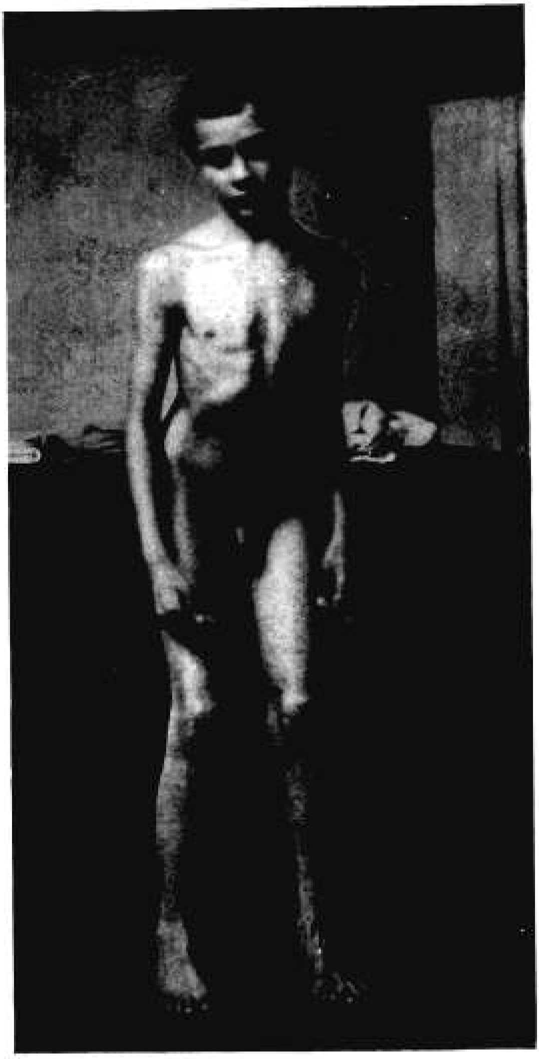

Fig. I.

Attitude of patient with Friedreich's disease. The feet are too far apart and the body is bent forward. The oscillation of the head prevented a clear photograph of it.

was difficult for her to stand still without swaying, and this swaying was increased by closure of the eyes. Her knee jerks were lost. There was considerable ataxia of the hands in voluntary movement, and in all the automatic acts of dressing, fixing the hair, etc., the ataxia was apparent. There appears to be no disturbance of sensation in any part of the body to touch, temperature or pain impressions. The irregularity in the action of the bladder is not constant but occasional. She frequently wets the bed at night. A slight scoliosis is present, dorsal with convexity to the left, but there is no evidence of Pott's disease. There is some unsteadiness of the muscles supporting the head, and in consequence peculiar nodding motions of the head are seen, both, held at rest and while walking. Her pupils react normally, both, to light and accommodation, and her optic discs are clear. Her mental condition appears to be good.

Case II. Paul K., thirteen years of age in January, 1898. He is the third of nine children, only three of whom are living, the others having died in infancy from diseases not of a nervous character. He has one sister older and one younger than himself, neither of whom is affected by the disease. He is, therefore, the only member of the family thus far affected. His father died of paresis one year ago. His mother has had several miscarriages. The probability of a syphilitic inheritance is therefore great. The boy has always been delicate, has had numerous children's diseases, has suffered for long periods from gastro-intestinal derangement, and at the age of two had a large abscess of the neck. He has always been considered weak and feeble, and was never bright at school, it being remarked that he laughed at everything in a silly manner, and was not able to learn quite as well as other children; yet he appears fairly intelligent, is able to read and write, and, though having a dull facial expression, is not, apparently, weak-minded. His present illness began at the age of ten, very gradually, it being noticed that he was awkward in his gait, would stumble in going up and down stairs, and at the same time became clumsy in the movements of his hands. About the same time his mother noticed peculiar nodding or oscillating movements of the head, a tendency to look downward a good deal of the time. She says that he has always wet the bed at night, but that this has become more frequent of late.

Examination showed a very marked ataxia of gait, it being impossible for him to walk a straight line. His body stoops somewhat forward, as shown in the photograph, and it is impossible for him to raise the toes or feet from the floor while standing on his heels. He lifts his feet too high in walking, his steps are of irregular length; there is a tendency of the knees to overlap, though this is corrected by voluntary efforts to preserve his equilibrium, the toes fall down and inward as the foot is raised from the ground, making it necessary to step high in walking, and any attempt to stand or walk with the eyes closed results in a fall. The feet are markedly misshapen, the instep being too high, the foot somewhat clubbed, and the great toe and all the toes to a less degree being overextended so that all the tendons stand out upon the back of the foot. This deformity is shown in the photograph, in which a normal foot of about the same size is shown for contrast. He is able to place the foot flat upon the floor which corrects to some extent the deformity, but does not affect the hyperextension of the toes. All voluntary movements of the feet and hands are extremely ataxic. There appears to be a tendency to hold the fingers in a flexed position, suggestive of a beginning claw-hand. There is no scoliosis. There is constant oscillation of the head of slight degree laterally and antero-posteriorly, and this oscillation is increased on efforts of walking. His knee jerks are very much exaggerated, but there is no clonus. There are no sensory disturbances to tests of touch, temperature or pain. Muscular sense is much impaired; there are no elbow

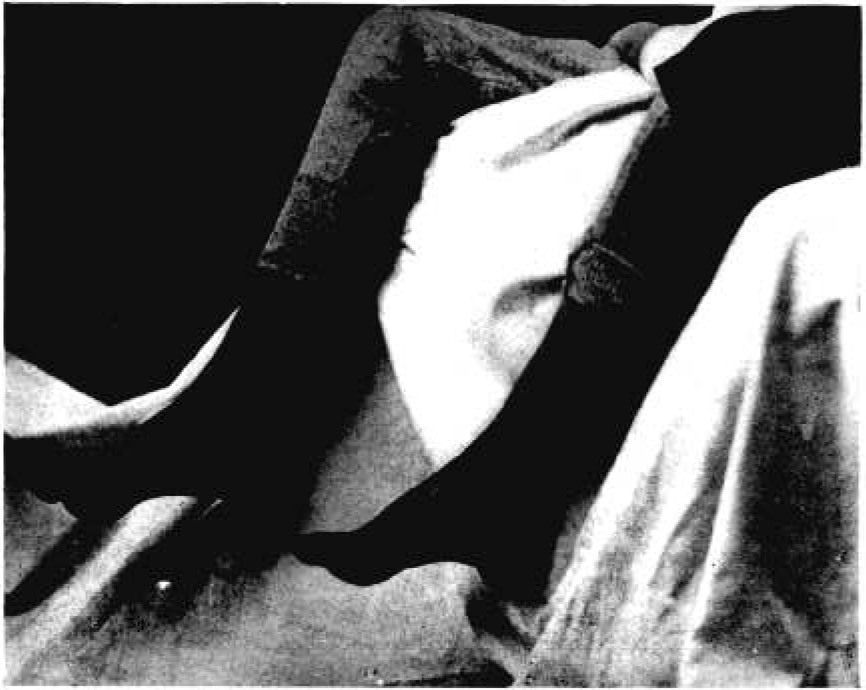

Fig. II.

The foot of the patient with Friedreich's disease contrasted with a normal foot. The high instep, the greater arch, and the retracted great toe are noticeable.

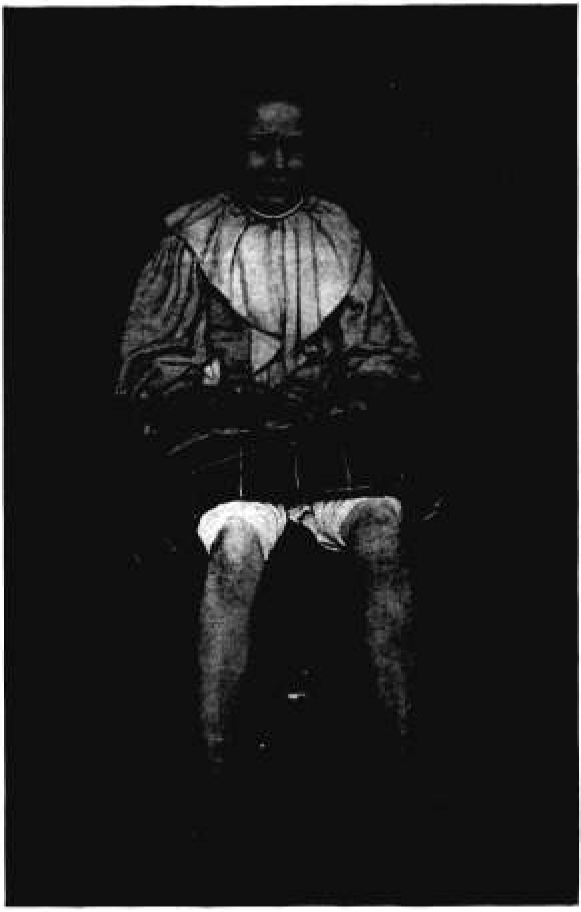

Fig. III.

Appearance of the legs and feet in Friedreich's disease. The slight ptosis is also visible.

or wrist jerks. His pupils react normally to light and accommodation. He has slight lateral nystagmus in both eyes on looking far to either side. Optic discs normal.

Case III. Female, aged eighteen in December, 1897. She is the only child in the family, and both parents are perfectly healthy, and there is no history of any similar affection to be found in any member of the family. Her disease developed about the age of six, after an attack of measles, and has been gradually progressing ever since, but its progress has been so slow that it is difficult to fix a date for the onset of special symptoms. It is known, however, that by the time she was thirteen, her walking was very difficult, and that she had by that time become awkward in her hands and unsteady in her head. Examination showed a rather thin, stupid-looking girl, with her head habitually held slightly forward and downward in a slight state of oscillation, which was increased by the act of walking. She seemed to have some difficulty in raising the eyes or in looking upward. This is evident in the photograph. There is no strabismus, but she has a slight lateral nystagmus on turning the eyes to the side, and the eyelids cannot be fully elevated, and the eyeballs cannot be rolled upward as far as normal. The pupils react to light and accommodation, and the optic discs are normal. She has marked ataxia of the hands on all movements, and very great ataxia of the legs in walking, so that the act is an irregular stepping one with characteristic deformity of the feet, tendency to drop foot with incurvation of the foot and bending forward of the entire body in the act of balance to a slight degree. She has a slight lateral curvature of the spine toward the right which has required the use of braces for about two years. She has no disturbance of the bladder or rectum and no sensory symptom of any kind. The knee jerks are lost. The awkwardness of her hands is very marked in the act of dressing. She appears to be dull mentally, takes very little interest in things about her, and is rather slow in her speech, with some hesitation. Her mother, however, will not admit that she is weak-minded in any degree.

These cases are grouped together as they are fairly typical of Friedreich's ataxia. It is interesting to notice that two of them began after an attack of measles. Considering the frequency of this disease in children and the fact that it often runs through a family, affecting several members, the question may be raised whether too much attention has not been given to the supposed hereditary factor in this disease. I have records of three other cases of Friedreich's ataxia, and in none have I been able to ascertain any evidence of similar disease in a previous generation. As the disease does not affect the duration of life, and the existence of a chronic cripple in a family cannot be overlooked or forgotten, it seems to me evident that many cases do not rest upon an hereditary basis. Nor can it be stated with precision that this is a disease of maldevelopment, for in the majority of cases children have grown to the age of six or eight without any manifestation of the disease, and have learned to walk well. The degenerative changes, therefore, in the nervous system must supervene upon healthy nervous tissue, and while it may be regarded as probable that from some unknown reason the life period of some neurons may be unusually short in these cases, just as the life period of muscles is abnormally short in cases of dystrophy, yet it is manifestly wrong to speak of a congenital maldevelopment in these cases. That the degenerative process is not exclusively limited to the spinal cord is manifest from the existence of ocular and mental symptoms in all the cases. Nystagmus was present in all, a tendency to ptosis and manifest weakness of the superior rectus muscle was present in two. Marked mental dullness was present in all three. It is, therefore, probable that the influence which leads to the disease is one which affects the entire nervous system, and if, as seems very probable, the occurrence of acute infectious diseases, notably measles, is the exciting cause, this disease may be in future classified with multiple sclerosis, as a nervous sequel of the infectious diseases of children.

The possibility of Marie's disease, in the second case, is admitted. As yet it seems impossible to draw a sharp line between Marie's disease and Friedreich's disease clinically.

77. SPLITTING THE KIDNEY CAPSULE FOR THE RELIEF OF NEPHRALGIA.

In the Medical News, Jan. 30th, 1897, Dr. George B. Johnson reports two cases in which violent pain, resembling attacks of renal colic, led to the operation of nephrotomy for supposed calculus, although the urinary symptoms were not charateristic. In neither case was stone found to be present. The kidneys, however, appeared to be too tightly enclosed in a tense capsule. In each case a free slit was made in the whole length of the kidney capsule. The pain was promptly abated by this procedure and never returned. The author concludes:

1. Nephralgia is not always associated with a demonstrable lesion.
2. When other evidences of kidney disease are wanting, the pain is perhaps due to a too-tight capsule.
3. Nephralgia may, and frequently does, simulate symptoms of gross tissue changes or presence of mechanical irritants.
4. When severe and persistent pain in the kidney exists, without other evidences of renal involvement, exploratory operation is indicated.
5. When inspection, palpation, and needle punctures fail to disclose a condition sufficient to account for the pain, the capsule should be freely opened.

SHIVELY.